Primary Infrapapillary Carcinoma of the Duodenum : Report of a Case

AUTHOR(S):

KASAHARA, YOH; YAMADA, YUKIKAZU; UEDA, SHOZO; NAKAO, KIICHI; SONOBE, NARUMI; TAKEMOTO, MASAHIKO; SUDO, TAKAAKI; UMEMURA, HIROYA; SHIRAHA, SEI; KUYAMA, TAKESHI

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Primary Infrapapillary Carcinoma of the Duodenum: Report of a Case

Yoh Kasahara, Yukikazu Yamada, Shozo Ueda, Kiichi Nakao, Narumi Sonobe, Masahiko Takemoto, Takaaki Sudo, Hiroya Umemura, Sei Shiraha and Takeshi Kuyama

The Second Department of Surgery, Kinki University School of Medicine
(Director: Prof. Dr. Takeshi Kuyama)
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Primary duodenal carcinoma (PDC) is a rare disease. In almost 500,000 autopsies reviewed by Kleinerman et al[9], the incidence of primary malignant neoplasms of the duodenum was 0.035 per cent. The incidence of carcinoma of the duodenum accounted for 0.04 per cent[14] to 0.06 per cent[12] of the autopsied cases, and 0.3 per cent[6] to 0.4 per cent[10] of all carcinomas of the gastrointestinal tract. Were carcinoma of the papilla of Vater excluded, the incidence of PDC would be perhaps very low. The location of PDC relative to that of papilla may play a role in determining treatment.

Report of a Case

A 71-year-old female complaining of pain in the RUQ of the abdomen of 7 month's duration was admitted to a municipal hospital on November. 1983 because of postprandial nausea and vomiting. She was transferred to our hospital on January 11, 1984. Family history revealed maternal rectal carcinoma. Personal history revealed appendectomy in her 30's and hypertension for the past 4 years. Physical examination showed an emaciated woman 151 cm in height, 38 kg body weight, blood pressure 100/60 mmHg, regular heart rate 68/min, with regular respiration. She had no jaundice, the conjunctivae were anemic, systolic murmurs were heard at the left 5th ICS, and a small node was present in the right groin. Abdominal inspection and palpation revealed a mass of bloated stomach in the upper abdomen and another fist-sized, smooth mass without tenderness or respiratory wandering on the right side of the navel.

Usual laboratory data showed slight anemia, moderate insufficiency of renal function and marked metabolic alkalosis (Table 1). Barium swallow revealed a mass in the second portion of the duodenum, without complete obstruction. Endoscopic inspection and biopsy of the mass revealed an irregular bleeding ulcer just beneath the papilla of Vater, with a histology of adenocarcinoma. Ultrasonography (US) of the abdomen showed a 4×3 cm mass adjacent to the head of the pancreas (Fig. 1). Selective celiac angiography was impossible because of the sclerotic

Key words: Primary duodenal carcinoma, Infrapapillary carcinoma, Pancreatoduodenectomy, Segmental excision, CUSA.
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Present address: The Second Department of Surgery, Kinki University School of Medicine, Sayama-cho, Osaka, 589 Japan.

Table 1. Preoperative laboratory data

RBC ($\times 10^4$)	311	Artery blood pH	7.515
Hb (g/dl)	10.0	SBE (mEq/l)	+9.6
Ht (%)	29.7	GOT (IU)	25
WBC	6,500	GPT (IU)	22
		Alkaline-P (IU)	70
Creatinine (mg/dl)	2.3	LAP (GU)	113
BUN (mg/dl)	50	Gamma GTP (mU/ml)	7
Na (mEq/l)	140	s-Amylase (IU)	459
K (mEq/l)	2.9	Elastase-I (ng/dl)	1,700
Cl (mEq/l)	74	CEA (ng/ml)	2.6
Creat. Cl. (ml/min)	21.0-44.7	Urinalysis	n.p.

tortuosity, but abdominal aortography showed partial irregularity of the postero-superior pancreato-duodenal artery without obvious change in the intrapancreatic arteries or the portal venous system. Cytology of the duodenal fluid was Pap. II. ^{67}Ga scintigraphy showed

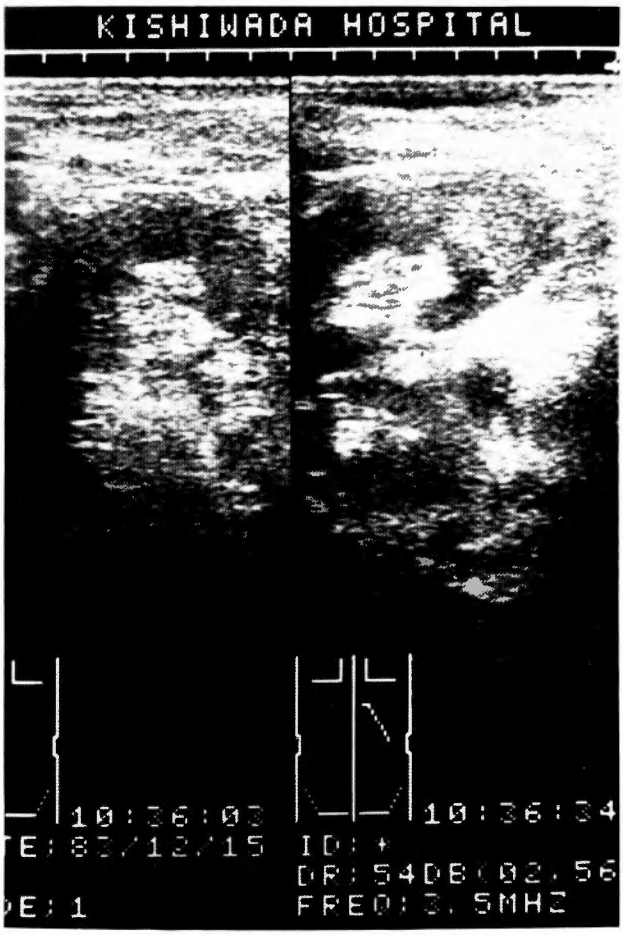

Fig. 1. Ultrasonogram of the abdomen showing a huge mass in the right upper abdomen.

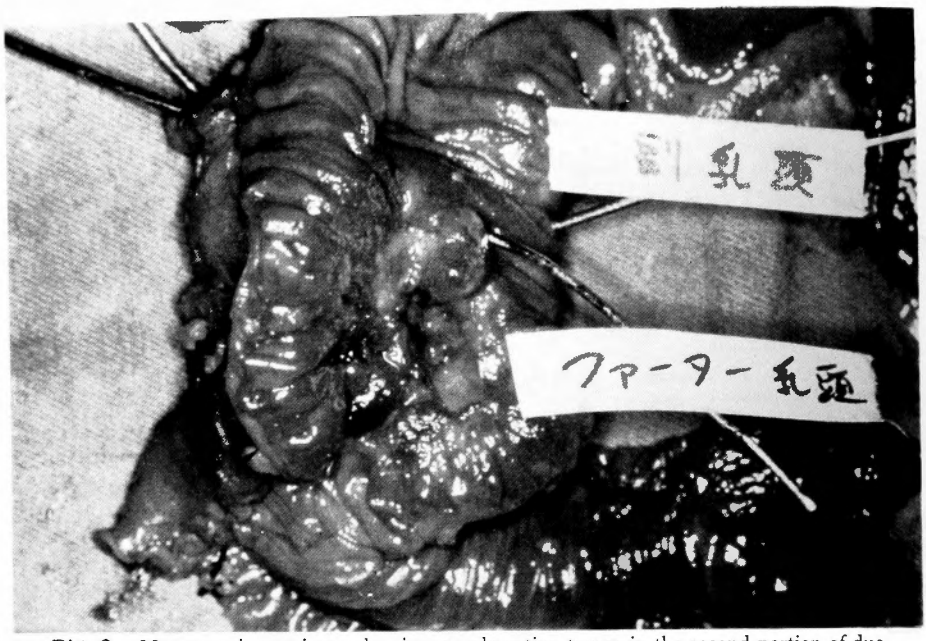

Fig. 2. Macroscopic specimen showing an ulcerative tumor in the second portion of duodenum; the passage through papillas are demonstrated by two probes.

abnormal accumulation in the region of the pancreatic head.

With a diagnosis of duodenal carcinoma, the patient underwent pancreatoduodenectomy on January 25, 1984. The tumor had invaded the capsule of the right kidney and a hard, massive metastasis was noted around the superior mesenteric vessels. To preserve renal function, right nephrectomy was avoided and the invasion was divided by CUSA (Cavitron; Ultrasonic Surgical Aspirator). The perivascular metastasis was also divided by CUSA. Anastomoses were done using a modified Child's method. Postoperative course was uneventful except for slight abnormalities exhibited in liver chemistry studies. Since discharge on March 12, 1984 she had been doing well. The resected specimen showed $7 \times 5.5 \times 4$ cm ulcerative tumor obstructing the second portion of the duodenum. The upper edge of the tumor was approximately 2 cm distal to the papilla of Vater, the passage through the papilla being intact (Fig. 2). Photomicroscopic examination revealed cancer nests with columnar epithelia partially invading the pancreatic tissue (Fig. 3). Tissues of the papillary region were normal. Postoperative diagnosis was primary carcinoma de novo from the duodenal mucosa.

Discussion

The location of duodenal carcinoma origin can be divided into three regions: suprapapillary, peripapillary and infrapapillary. However, so-called carcinoma of the papilla of Vater includes cancer of the bile and pancreatic ducts, in addition to true cancer of the duodenal mucosa, and differentiation of these cancers in the advanced stage is very difficult both macro- and microscopically. Recently carcinoma of the peripapillary region has been excluded from the primary

Fig. 3. Photomicroscopic specimen of the carcinoma (H.E. stain, ×100).

duodenal carcinoma group, excepting precisely diagnosed cases of duodenal mucosal origin[13].

The duodenum, though only 30 cm long in adults, is divided into four parts anatomically; from the surgical viewpoint, however, it may be divided into the suprapapillary and infrapapillary regions. In our survey, carcinomas in the suprapapillary region were subdivided into cancer of the duodenal bulb (Bulb) and cancer of the other suprapapillary portions (Other-SP). Infrapapillary region carcinomas were subdivided into cancer of the second portion (Second) and cancer of the third portion distal to the duodenojejunal flexure (Distal).

In our review of 271 cases of PDC reported in the Japanese literature (including our own case) since 1970 (carcinomas of the papilla of Vater excluded), 155 cases of suprapapillary carcinoma showed a male-to-female ratio of 1 : 1, with average ages of 64.4 years in males and 61.9 years in females; 99 cases of infrapapillary carcinoma showed a ratio of 1.9 : 1, with average ages of 54.6 years and 54.3 years. Patient distribution by PDC site is shown in Table 2. Elderly patients were prevalent in the suprapapillary carcinoma category. Tke prevalence of suprapapillary over infrapapillary carcinoma (1.7 : 1 in our survey) was reported in other authors[4,13] as well, except for JOESTING et al[7].

Although the first report of PDC in Japan was made by HAYASHIKAWA and KANAMORI[5] in 1898, only reports since 1970 were reviewed here, since the remarkable development in the US in the '70's of such diagnostic techniques as hypotonic duodenography, duodenoscopic examination with biopsy and clinical imaging have made reports from that time more reliable in detail than those previous. Almost all of the PDC in our survey developed de novo from the duodenal mucosa. Malignant alteration of benign adenoma, ectopic pancreas or carcinoid are exceptional origins of PDC. Recently MORI et al[11] stated that more than half of the early cases of PDC

Table 2. Relationship among the age, sex and location of PDC

Age of yrs.	Suprapapillary				Infrapapillary					
	Bulb		Other-SP		Second		Distal		Undescribed	
	M.	F.	M.	F.	M.	F.	M.	F.	M.	F.
80's	3		3							
70's	8	10	13	11	3	1	4	2		1
60's	5	5	17	19	2	2	11	7	1	
50's	3	7	16	13	4	2	9	6	2	
40's	1	2	5	4	4	2	8	4		
30's		2	2	1	3		1	1		
20's					1		2	3		

Other-SP means carcinomas excluding carcinoma of the bulb, Distal means carcinomas from the third portion distal to the duodeno-jejunal flexure, M. means male and F. means female.

showed malignant alteration of the villous adenoma.

Although the usual symptoms of upper gastrointestinal tract obstruction, gastrointestinal bleeding with or without resultant anemia, abdominal pain, loss of body weight, jaundice and/or palpable abdominal mass were main signs and symptoms of PDC, all were not pathognomonic. In the survey of BRENNER and BROWN[2], symptoms mainly of gastrointestinal or biliary tract obstruction, peptic ulcer and bleeding were noted. Nausea, vomiting and/or abdominal distention as obstruction symptoms were noted in 130 of 251 patients (51.8%) in our survey. Similarly, hematemesis, melena, occult fecal blood and/or anemia with bleeding symptoms were noted in 105 patients (41.8%), abdominal pain in 88 (35.1%) and palpable abdominal mass in 28 (11.2%). The lower incidence of jaundice (28 patients, 11.2%) may reflect the strict elimination of carcinoma of the papilla of Vater. Seventeen asymptomatic patients were incidentally found during mass screening for gastric cancer or gastroduodenoscopy for vague abdominal complaints. Almost all had early suprapapillary carcinoma.

Table 3. Relationship between the signs and symptoms and location of PDC

	Suprapapillary				Infrapapillary			
	Bulb		Other-SP		Second		Distal	
	No.	(%)	No.	(%)	No.	(%)	No.	(%)
Nausea and/or vomiting	12	26.1	40	39.2	14	56.0	35	67.3
Abdominal distention	9	19.6	18	17.6	8	32.0	14	25.9
Total*	18	39.1	45	44.1	14	56.0	39	72.2
Bleeding	11	23.9	28	27.5	8	32.0	19	35.2
Anemia	5	10.9	33	32.4	9	36.0	17	31.5
Total*	12	26.1	43	42.2	13	52.0	25	46.3

The percentage was calculated in 46 patients of Bulb, 102 of Other-SP, 25 of Second and 54 of Distal.

* The number of total excluded cases of duplicated symptoms.

The incidences of obstruction and bleeding symptoms in PDC in a portion of the duodenum are listed in Table 3. Obstructions increased as the PDC became more distal. Bleeding was less common in cancer of Bulb with almost the same frequency in the other three portions. SPIRA et al[15] state that the signs and symptoms of PDC varied with the location of the tumor within the duodenum and also with the gross characteristics of the tumor itself. BURGERMAN et al[3] classified the macroscopic appearance of PDC into the four types; 1) ulcerative, 2) polypoid, 3) annular constrictive and 4) diffuse infiltrative. The third and fourth types, rare, were omitted in our survey. The relationships among symptoms, locations and macroscopic types are shown in Table 4. The appearance of symptoms was less frequent in the polypoid type, which, together with the ulcerative type, showed equal frequency in cancer of Distal. Carcinoma of the polypoid type was prevalent in the Bulb. Carcinomas in the other portions were prevalently of the ulcerative type.

Although the gastrointestinal series, computed tomography, US and selective angiography may play an important role in the assessment of extension, resectability and so on, accurate preoperative diagnosis of PDC is established by duodenoscopic speculation and biopsy. More than 60 per cent of the patients in this study were confirmed as to PDC before surgery or autopsy. Although diagnostic difficulty may increase when the PDC is located distally, our survey revealed no significant difference in the ratio of precise preoperative diagnosis in an portions of the duodenum.

Differential diagnosis is difficult in the advanced PDC in the second portion. So-called carcinoma of the papilla of Vater, carcinoma of the pancreas head or of the lower bile duct should be eliminated. In far advanced cases, differential diagnosis among these carcinomas may be impossible at surgery and at histological evaluation. Carcinoma of the Bulb, especially in the ulcerative type, has often been misdiagnosed. Eight cases in our survey had ulceration or penetration resembling a benign duodenal ulcer.

Treatments involving endoscopic maneuvering such as polypectomy, infusion of cytotoxic agents or cauterization by laser were reported in several cases whose surgical indications were poor. Generally, however, surgical removal is the PDC treatment of choice, surgical procedures

Table 4. Relationship among the signs and symptoms, macroscopic type and location of PDC

		Suprapapillary				Infrapapillary			
		Bulb		Other-SP		Second		Distal	
		No.	(%)	No.	(%)	No.	(%)	No.	(%)
Obstruction	U.	7	41.2	20	41.7	6	46.2	20	71.4
	P.	7	24.1	13	56.5	1	12.5	9	75.0
Bleeding	U.	5	29.4	27	56.3	7	53.8	13	4..4
	P.	5	17.2	7	30.4	3	37.5	6	50.0

U. means ulcerative type; This type was found in 17 patients of Bulb, 48 of Other-SP, 13 of Second and 28 of Distal.
P. means polypoid type; this type was found in 29 patients of Bulb, 23 of Other-SP, 8 of Second and 12 of Distal.

Table 5. Relationship among the macroscopic type, surgical procedure and location of PDC

	Suprapapillary				Infrapapillary			
	Bulb		Other-SP		Second		Distal	
	U.	P.	U.	P.	U.	P.	U.	P.
Segmental or local excision	8	18	3	12	2		14	5
Pancreato-duodenectomy	4	5	33	6	10	5	6	1
Anastomosis only			2		1	1	2	5

differing with site, degree of extension and so on. In Table 5, relationships among the PDC site, type and surgical procedure are listed. Segmental or local excision was preferred in carcinomas of the Bulb and Distal. For carcinomas of the second portion, regardless of the papillary position, pancreatoduodenectomy was indicated, whereas several polypoid carcinomas of Other-SP offered opportunities for segmental or local excision. In general, surgery was less invasive in polypoid than in ulcerative carcinoma.

The interval between onset of PDC symptoms and surgical intervention were given for only 109 patients (mean interval: 5.5 months) in our review. KERREMAN et al[8] gave a similar interval (6.9 months, mean time) for 12 patients. ALWMARK et al[1] reported a mean time of 7.5 months from first outpatient visit to surgery. Among the surgical procedures, 43 cases of segmental excision or local resection had a mean time of 4.1 months from onset of symptoms to surgery, 50 cases of pancreatoduodenectomy had a mean time of 6.0 months and 15 cases of anastomosis only the figure was 7.7 months. Although duration of symptoms may be an important factor in PDC surgery, five cases of carcinoma of the Second, whose duration of symptoms had a mean length of 2.5 months, were unresectable.

PDC sizes are shown in Table 6. The mean size was 4.9 cm in 57 patients who underwent pancreatoduodenectomy and 3.6 cm in 44 patients undergoing segmental excision or local resection. The surgical strategy may depend upon the site, macroscopic appearance, duration of symptoms, extension and size of PDC. JOESTING et al[7] demonstrated that survival in PDC was directly related to nodal metastasis and to grade of lesion in their collective review of 104 patients in the Mayo Clinic They also concluded that segmental excision was the preferable operation for infrapapillary carcinoma in selected cases, the 5-year survival rate for 46 patients who underwent radical resection for PDC being 45.7 per cent. Node metastasis, direct invasion

Table 6. The sizes of PDC (maximum length of the resected specimen)

	Suprapapillary				Infrapapillary			
	Bulb		Other-SP		Second		Distal	
	U.	P.	U.	P.	U.	P.	U.	P.
Less than 5 cm	7	19	18	12	5	5	11	4
5–10 cm	4	4	20	8	6		9	3

of adjacent tissues and/or distant metastasis were noted in 6 patients of carcinoma of the Bulb (12.8%), 28 patients of the Other-SP (25.9%), 11 of the Second (44.0%) and 25 of the Distal (45.5%), respectively, in our survey. If nodal state is a decisive factor in prognosis, the prevalence of nodal metastasis in the infrapapillary carcinoma seems to go counter to the results of ALWMARK et al[1] that the infrapapillary carcinoma had a better prognosis than the suprapapillary type. As to establishing prognosis, however, the small number (three patients[16,18]) who achieved 5 year survival in our survey was not enough upon which base a discussion. The mortality during follow-up was 68 out of 156 patients (43.6%) who underwent surgery.

Although adjacent extension of PDC is rapid and nodal metastasis is frequent, distant metastasis is said to be rare[17]. In order to relieve obstruction and/or bleeding symptoms, if palliative, PDC resection should be considered in advanced stage cases. CUSA is thought to be one of the useful devices for this. The increasing incidence of early PDC (33 patients in our survey), a result of mass screening for gastric cancer or gastroduodenoscopy for vague abdominal complaints, may gradually ease the formidable prognosis[3,15] of PDC.

Summary

A case of infrapapillary PDC in the second portion is presented. Although the patient had advanced carcinoma with invasion into the right renal capsule and other sites, palliative pancreatoduodenectomy was performed successfully with using a CUSA. In our review of 271 cases from the Japanese literature since 1970, several conclusions can be drawn regarding infrapapillary PDC in the second portion compared with that in other portions. Second portion PDC has: 1) the lowest occurrence rate; 2) the highest male-to-female ratio (2.4 : 1); 3) a prevalence of ulcerative over polypoid type (1.6 : 1); 4) greater frequency of obstructive symptoms than does proximal PDC; 5) the highest incidence of bleeding symptoms and 6) pancreatoduodenectomy as almost the only reasonable treatment. (Portions of this report were delivered at the 136th meeting of the Kinki Surgical Society on October 20, 1984 at Ohtsu City.)

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和文抄録

原発性十二指腸乳頭下部癌の1例

近畿大学医学部第二外科（主任：久山 健教授）
笠原 洋，山田 幸和，上田 省三，中尾 稔一，園部 鳴海
竹本 雅彦，須藤 峻章，梅村 博也
白羽 誠，久山 健

狭窄症状を呈した71歳女子の1例に対して CUSA を用いて膵頭十二指腸切除を施行し，良好な術後経過であった．切除標本で癌は Vater 乳頭下縁から約 2 cm 離れた部分が上限で，同乳頭の開存は保たれていた．Vater 乳頭部癌を除いての原発性十二指腸癌の頻度は極めて少なく，その中でも乳頭下部癌として十二指腸第二部に生ずるものは稀である．進行十二指腸癌の場合でも，愁訴除去のためには出来得る限り切除手術を行うべきと思われた．1970年以降の本邦における原発性十二指腸癌（乳頭部癌を除く）271例について考察し，特に本例のような第二部の乳頭下部癌について得た若干の知見について述べた．